ACUTE CIRCUMSCRIBED ŒDEMA.

N. O. M. Surg. Jnl, Sept 1887. vol 15. pp. 257-264

By Rudolph Matas, M. D.

Read before the Orleans Parish Medical Society.

It was in the month of May, I believe, in 1886, when I was called to attend a Mrs. V. G., an Italian woman, aged thirty-six, who complained to me of the symptoms which constitute the basis of this narrative. Before proceeding with the details of her case, however, I will state that she had been my client several years previously, during which time I had on several occasions attended to her husband and her children (five in number) for various ailments, among which, however, predominated the disorders due to malarial causation. The patient herself enjoys an average good health, though she somewhat exemplifies, in her dark, sallow complexion, etc., the "tropic diathesis" so ably described by our distinguished fellow member, Prof. Elliott, and which is so commonly met with among the laboring classes of our Italian population. In justice to her health, however, I must state that, outside of malarial fevers and a few accidental complications of childbirth, she rarely suffered with disease prior to the occasion when she called me to relieve her of the condition now under consideration. When I reached her house, which was late in the evening of the day that I had been sent for, I found her apparently as well as I had ever seen her and attending to her usual avocations. As I made some remark to the effect that she appeared too well to require medical assistance, and expressed some surprise at her sending for me, she told me that if I had only called a few hours earlier I would not have thought that she looked so well. She was feeling now, as usual in the evening, quite well; but it was "quite a different thing in the morning." In fact, for the last two months and over, she had been afflicted with a peculiar disfigurement of the face, which "could hardly be designated as sickness," for it

seemed, to her, too much of a trifle in the beginning to be worthy of such a name, but now she was becoming alarmed as it persisted so obstinately and prevented her, whilst it occurred, from attending to outside work on account of the distortion of her features that it entailed.

This "disfigurement," she explained to me, was a large swelling that took place daily between 8 and 11 A. M., which appeared in the upper lip and caused it to rise to the level of her nostrils, obstructing the nares and projecting over two and three inches beyond the surface of the teeth. The swelling came on gradually, but continued to increase for some hours until it reached a maximum about 12 M. or 1 P. M., when it gradually subsided, so that by three, four or five in the evening it had totally disappeared, leaving no trace behind it.

The swelling was perfectly painless, and to use her expression was "doughy" to the feel, and only bothered her by its ungainliness.

I must state that I was at once much struck by the singularity of the phenomenon, and especially by the clearness and accuracy with which the signs and symptoms were detailed to me. The periodicity of the affection and its long duration, above all, were curious. The swelling "came on regularly every morning and disappeared every evening; sometimes, though rarely, it appeared in the evening and disappeared late at night," was the answer always given by the patient when interrogated on this point; and other women, relatives and neighbors, who were about, all substantiated the statements by their repeated reiterations. I asked if any chill accompanied the appearance of the swelling, if it was followed or associated with fever or any other general disturbance, and to these queries the patient always gave a negative answer, stating, in addition, "that she was not prevented from doing any work about the house by the presence of the enlargement, and it was only by the inconvenience of its appearance that she did not venture out of the house while it lasted."

I then examined her mouth for subgingival abscess,

periostitis, dental caries, or other mouth lesions which so commonly cause peri-oral swellings, but I failed to see any lesion that could reasonably account for the swelling of the upper lip, though some of the molars were carious. The absence of any inflammatory lesion about the lips *at the time* the examination was made would also exonerate the teeth from any participation in the trouble, as œdemas of dental or gingival origin are not so transitory in character; and, furthermore, are not known to oscillate in such rythmical fluctuations as this had done for so long a time as three months. What, then, could cause such a periodic and painless œdema, as I at once characterized the condition? Could it be an occult renal, cardiac or hepatic disease with albuminuria? The case puzzled and interested me. Without prescribing I requested the patient to send me some of her urine the next day, and after examining her heart, which I found perfectly normal, and searching for other evidences of renal or hepatic disease, such as œdemas in other localities, etc., I concluded that this trouble was not associated with disease of any of these organs, though I relied on the examination of the urine for a final conclusion on the question of renal disease, which all that time struck me as the most probable morbid state in which such an œdema could arise. The next day the urine was carefully examined for albumen, casts and sugar, and the specific gravity carefully taken, and I distinctly remember that I found it normal in every respect. There was no anæmia, no evidence of any dyscrasia that could be incriminated; nothing, in short, that an attentive clinical survey could seize as an efficient factor in the causation of the trouble.

I decided to see the patient myself during the œdematous period, and that very morning, before 12 M., I called at the patient's house and was at once convinced of the truth of her assertions, by the remarkable alteration in the outlines of her face. Her lips, and especially the upper one, are thin normally and of a darkish red hue, but now the upper one projected from her face like a proboscis. The lip had

indeed assumed elephantine proportions and impressed on the features a curious, grotesque appearance. The enlarged labium hung over the lower like an overhanging tumor, of a pale, waxy, leaden hue, especially on its mucous portion. On touching the mass it felt, as the patient said, "doughy," pitting on pressure and feeling cold and almost clammy to the touch. The cheeks did not appear to participate in the infiltration and neither did the eyelids, the swelling appearing to blend gradually with the remainder of the face on a level with the naso-labial furrow.

I again examined the mouth to reassure myself, and took the temperature under the tongue, but the whole examination gave only negative results.

It was now time to prescribe, as the patient anxiously awaited for a remedy that would relieve her of her annoying condition, or for an opinion that would enlighten her on the prognosis of her case, the demand for a diagnosis being thus embarrassingly thrust before me.

If there was a striking feature in this case it was certainly the quotidian periodicity of the phenomenon, the rythmical recurrence of the œdema; and if there is a valuable clinical criterion of malaria—though not an infallible one—it is periodicity. Now the word malaria implies usually quinine—therapeutically, at least at the bedside, though, I must state, that the converse also appears to hold as well, if not better, with many—*i. e.*, that quinine implies malaria, or rather that the test of malaria is quinine. And surely no one long engaged in clinical work will gainsay that this test is often regarded as the crucial reagent for the detection of malaria.

But I will not digress at this moment. I will state simply that in the presence of this perplexing case the periodicity of the affection suggested malaria, and the malaria quinine; and with this suggestion I was at least temporarily armed with a diagnosis and therapeutic indication. I therefore ordered quinia sulphate, twenty grains, in capsules, to be administered in the evening, and ten more early in the morning, at least three hours before the usual appearance of the œdema.

Two days after, at 2 P. M., I was much surprised to see the patient herself at my office, and what was most satisfactory was that she bore no evidence on her face of the annoying trouble, even to the presence of the heretofore hidrophilic labium. The patient was delighted that the œdema had not returned; she said she had been cured and had not delayed in coming to my office to apprise me of the news. I could hardly believe that the ailment had radically disappeared, and told her that it might return, but to guard against a further recurrence of the trouble, and not without some curiosity to know the ultimate results, I prescribed a capsule containing quinia and arsenic as a basis, to be taken three times daily. The next day I learned the patient had had a relapse, but not so well characterized as formerly. The tonic pills were then reinforced by the administration of twenty grains of quinine in the evening. After this the swelling did not return, and, as I have had occasion to see the patient quite recently, I have learned that she has been completely free from any return of the trouble since the time when I prescribed for her.

Such a well marked interruption in the course of disease, and its complete arrest following immediately upon the administration of remedy, could hardly be regarded as a coincidence, and in this I am convinced that the cessation of the œdema was entirely the result of the therapeutic action of the quinine. This practical result, together with the peculiar features of the case, did not fail to impress it in my memory. In the lapse of time it became gradually blended with my malarial recollections.

It was barely passing into the realm of reminiscence, however, when the details of the case were again roused to vividness by reading abstracts of two interesting communications made at the meetings of June 3 and 25, 1887, of the Imperial Royal Medical Society of Vienna, on the subjects of acute circumscribed œdema, by Dr. Riehl. In the first of these meetings the report says (*Semaine Medicale**) that Mr. Riehl presented two patients afflicted with acute cir-

**Semaine Medicale. Paris*, Nos. 23, 26. 1887.

cumscribed œdema of the skin. "This affection, which has been described by Quincke, consists in the sudden appearance and without any appreciable cause of an œdematous tumefaction, which, in the course of two or three hours, becomes very pronounced, and disappears completely at the end of twenty-four or thirty-six hours. This tumefaction is localized ordinarily in the dorsal aspect of the hand and in the face, in the eyelids, about the mouth, the lips, the cheeks, and sometimes even in the pharynx and larynx, where it is liable to produce suffocation." In one of Dr. Riehl's cases the affection has persisted for three years in the zygomatic region; in the other thirteen years in various parts of the body. At the meeting of the same society, held June 25th, Riehl presented further details in regard to these two patients. As to the last instance, that of the patient who became ill with the disease in 1874, he tells us that he is a man aged forty-one years, who had always been healthy prior to 1874, when, shortly after the death of his wife, he was suddenly seized with œdema in the left eyelid, which, appearing suddenly at night, disappeared as quickly the next morning. From this time on and at various intervals he presented œdematous localizations in various regions of the face. At present, the œdematous seizures are repeated every eight or ten days, which, outside of the inconvenience resulting from the altered appearance, cause no trouble.

In the other case he referred to a man thirty-three years of age, who was attacked three years before, eight days after his wedding, by an acute œdema of the left cheek. Since that time he has these limited œdemas every fifteen days.

In these two cases the general condition of the patient is all that could be desired. The urine is normal and there exist no hereditary antecedents. But this is not always the course of this affection. In a case reported by Quincke the appearance of the œdema was always associated with a general *malaise*, somnolence, vomiting, etc.

Strübing mentions three cases which were characterized

by intense gastric disturbance, and vomiting as often as twenty or thirty times daily, constipation, psychical disturbances, etc., even when the œdematous areas were insignificant in size. Hereditary influences appear to play a part in this affection, and *apropos* of this Strübing relates the case of an old man of seventy who was affected with acute circumscribed œdema, whose son suffered also with the paternal affliction. Analogous examples have been reported by Quincke, Falcone, etc.

The differential diagnosis, according to Rhiel, is based on the suddenness with which the œdema appears and in the absence of all inflammatory symptoms; the absence of albumen in the urine or any other evidence that would indicate a renal or other lesion that might otherwise reasonably account for it. The differential diagnosis between the acute circumscribed œdema and myxœdema should present no difficulties to the clinician.

The etiology of the disease is still obscure. The influence of traumatisms has been appealed to to explain the condition, and, in fact, Quincke succeeded in producing an acute circumscribed œdema in a predisposed subject, by inflicting slight blows on the face, etc., but this mechanical causation must entirely play a secondary role, acting purely as an exciting agent in already predisposed subjects. Moral disturbances have been regarded, and perhaps with some foundation, as important predisposing factors.

As to the actual phenomenon itself—*i. e.*, the œdema—it is accounted for by Quincke, for instance, as an angioneurosis. Strübing attributes it to an exaggerated excitability of the vaso-dilator nerves. Riehl opines that the difficulty lies much more in the central nervous mechanism and in its peripheral dependencies, and in support of this opinion he appeals to the migratory or metastatic character of these œdemas, and their association with nervous disturbances of a peripheral character and vomiting.

From the preceeding synopsis of Riehl's paper, which embraces the leading points presented by Quincke (in his

article in *Monatscheft für Practische Dermatologie*, No. 5, 1885) it will be seen that, outside of the leading clinical features of the affection, nothing very definite is known as to its real nature, notwithstanding the fact that three theories have been advanced to explain its main and characteristic phenomenon, the acute circumscribed (painless) œdema.